

RADIOLOGY CORNER

Echinococcosis of the Liver

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Note: This is the full text version of the radiology corner question published in the February 2007 issue, with the abbreviated answer in the March 2007 issue.

Echinococcosis is a parasitic infection that poses a potential health threat to military members and others who travel to endemic regions. While canines are the primary host of the *Echinococcus* tapeworm, sheep and humans serve as secondary or intermediate hosts. Human infestation begins following accidental ingestion of the *Echinococcus* ova via a contaminated food source. Following ingestion, the ova enter the blood stream or lymphatic system through the small bowel and eventually lodge in the parenchyma of the liver or lungs and then develop into cystic lesions that slowly increase in size over time. Imaging is helpful in identifying and characterizing the size, number and location of these lesions, thereby providing useful information for preoperative planning. Successful treatment usually involves a combination of surgical and medical therapy.

Introduction

Echinococcosis, although relatively uncommon in the United States, is a global public health problem. It remains a persistent concern in most endemic areas, posing a potential threat to military members who deploy to these rural and often economically depressed regions. The cystic lesions that characterize this zoonotic disease can be successfully treated following an appropriate clinical and imaging work-up. It is therefore important for military clinicians and radiologists to be familiar with the characteristic imaging features and clinical manifestations of this parasitic disease. The following case report reviews the typical clinical presentation, imaging findings, and treatment of Echinococcosis.

History

A 40-year-old male presented with upper abdominal pain and fever. Physical examination was noncontributory. An upper abdominal ultrasound revealed a large, nonspecific cystic lesion of uncertain etiology and demonstrated no evidence of acute cholecystitis. Further evaluation with contrast-enhanced MR imaging was performed (Figs. 1a-c).

Summary of Imaging Findings

T1-weighted (figure 1a), T2-weighted (figure 1b), and contrast enhanced T1-weighted MR imaging (figure 1c) of the liver shows two large predominately cystic right hepatic lobe lesions. The more superiorly located lesion contains at its posterior aspect, a smaller cyst, which has been described as a "daughter cyst" in the setting of Hydatid disease (see figure 1b).

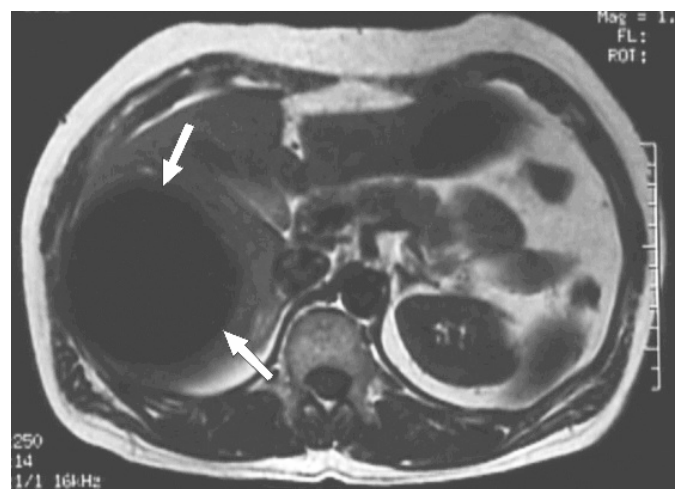


Figure 1a. Axial T1-weighted MR image through the liver and pancreas shows a well-defined, rounded large right hepatic lobe mass (arrows) demonstrating homogeneous very low signal intensity.

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Cyst aspiration was not attempted due to the concern for dissemination of the infected contents of the hepatic cystic lesions. Serologic testing was consistent with *Echinococcus* infection.

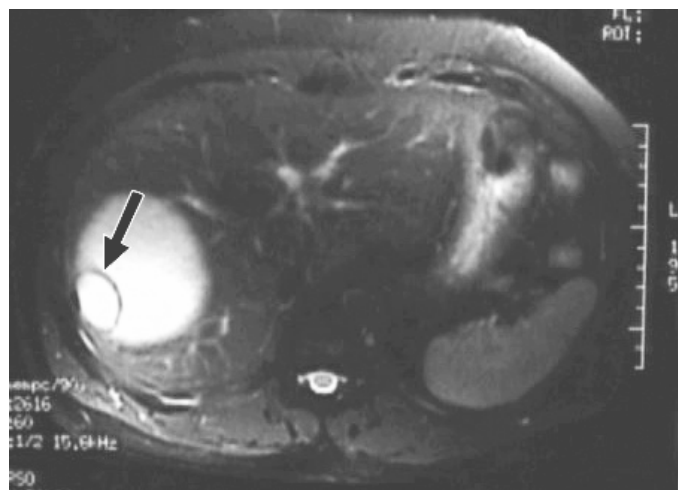


Figure 1b. Axial fat-suppressed T2-weighted MR image through the liver and spleen shows that the large rounded structure is predominantly cystic, but also reveals an additional smaller thin-walled oval cyst (arrow) within the larger cystic structure.



Figure 1c. Coronal fat-suppressed T1-weighted post-gadolinium MR image of the upper abdomen at the level of the aorta shows two large comparably sized hepatic cystic lesions. The more superiorly located cystic lesion demonstrates a thicker ill-defined wall (long arrow), while the cyst located in the inferior right hepatic lobe demonstrates a well-defined thin wall (short arrow). This second lesion corresponds with the lesion shown in figures 1a and 1b. The lack of internal enhancement confirms the cystic nature of these lesions.

Discussion

Echinococcosis, also known as Hydatid disease, is a tissue infection caused by the *Echinococcus* tapeworm in its larval stage. This zoonosis is endemic in rural, economically depressed areas where sheep and cattle are raised. The primary endemic areas are located in Australia, the Mediterranean,

South America, the Middle East, Africa, and Central Asia. In the United States, Echinococcosis is uncommon, but has been reported in New Mexico, Arizona, Utah, and California. The various species of *Echinococcus* include *E. granulosa*, *E. multilocularis*, *E. alveolaris*, and *E. vogeli*. The unilocular cystic form is caused by *E. granulosa* while the less common multilocular form is caused by *E. multilocularis* (1). Risk factors for contracting this infection are exposure to fecal material of infected canines, cattle, sheep, pigs, or deer.

The life cycle of the *Echinococcus* tapeworm involves canines as the primary or definitive hosts and sheep, cows, rodents, and humans as the secondary or intermediate hosts. *Echinococcus* infestation of the small intestine of the canine primary hosts results in the contamination of the fecal material with *Echinococcus* ova. The secondary host becomes infected following accidental ingestion of ova via contaminated food or water sources. Gastrointestinal enzymes are then responsible for breakdown of the external shell of the ova, thereby releasing the embryos. Following absorption by the intestinal mucosa, the embryos travel throughout the body via the venules and lymphatics and eventually lodge in various organs and tissues of the body. Next, the embryos develop into small cystic lesions that slowly increase in size over time. Each cystic lesion contains three distinct layers: the outer, pericyst; the middle, ectocyst; and the inner, germinal layer (1). The Liver is the most common site of larvae deposition (75%), while the pulmonary parenchyma is the second most common site of infection. Other locations of potential larvae deposition include the mediastinum, heart, brain, skeleton, muscles, spleen, and kidneys. Cysts can remain asymptomatic for extended periods of up to 20 years before they reach sufficient size to produce symptoms.

Clinical manifestations are relatively nonspecific and may include: upper abdominal pain, fever, cough, chest pain, bloody sputum, and pruritis. Although small cysts may be asymptomatic or unrecognized for many years, they have the potential to develop into extremely large lesions, some reportedly the size of a basketball (2). When large, a single hydatid cyst may prove fatal. Depending upon the species of organism, the cysts may be unilocular or multilocular. Each hydatid cyst contains numerous protoscolices (ranging from hundreds to thousands per cyst), each with the potential to develop into an adult worm following ingestion by a canine host and each with the ability to develop into a separate hydatid cyst if the “mother” hydatid cyst ruptures.

Ultrasound, CT and MRI are all appropriate for imaging of Echinococcal liver disease. Imaging early in the disease process often reveals one or more simple appearing cysts of varying size that are surrounded by either a thin well-circumscribed wall or less often a by a slightly thickened irregular wall, with the wall thickness ranging from 1 to 10 mm. The next stage of the life cycle involves the development of “daughter cysts” (3). At this stage, the cyst will typically demonstrate low echogenicity on ultrasound, hypodensity on CT, and low to intermediate signal intensity on T1- and high signal intensity on T2-weighted MR images. During the larval stage, the scolices begin to separate from the germinal layer and fall to the dependent portion of the cyst, appearing on

imaging studies as internal debris that is referred to as “hydatid sand” (4). High internal pressures can lead to disruption of the internal membranes and give the appearance of an undulating internal membrane. This imaging finding is nearly pathognomonic for hydatid disease and can be seen on ultrasound, CT, or MR imaging (1). Peripheral calcifications infrequently develop in the later stages of cyst development and when present are best depicted on noncontrast CT images.

The pulmonary parenchyma is the second most common site of involvement in adults and the most common site of involvement in children (2), and within the lungs, there is a predilection for involvement of the lower lobes. The lesions are typically first detected on chest radiography and often further characterized by CT imaging, while MR imaging is rarely necessary in the evaluation of pulmonary Echinococcus. On chest radiography the Echinococcal lesions can present as either a solid appearing mass or as an air filled cystic structure. Communication of the outer layer of the cyst with the bronchial tree can create a thin crescent of air within the cyst, referred to as the crescent or meniscus sign. Pulmonary lesions rarely calcify or develop “daughter” cysts.

The “Water-Lily” sign refers to a collapse of the endocyst layer that results in the inner cyst lining falling into the fluid in the dependent aspect of the cystic lesion. This gives the appearance of debris floating on a layer of fluid within the cyst (1). An empty cyst can occur if the cyst contents are expectorated. In the lungs, multiplicity occurs in up to 20% of patients. A potentially serious complication of pulmonary involvement is the rupture of a cyst with spillage of the contents into the airways or pleural space. In up to one-third of patients this complication has already occurred at the time of initial presentation.

Following treatment, hydatid cysts of the liver are best followed sonographically due to the absence of ionizing radiation and ease of access to this modality. The cysts will gradually fragment, collapse, and reduce in size leading to the sonographic appearance of a pseudosolid or echogenic mass. At this stage, the lesions can easily be mistaken for a tumor or abscess. Color Doppler can help differentiate a collapsed Hydatid cyst from a tumor as the collapsed cyst will demonstrate lack of internal vascularity, while a tumor in the absence of necrosis will usually demonstrate internal blood flow. Differentiation of this echogenic appearance from an abscess usually requires clinical correlation.

A definitive diagnosis can be made with serodiagnostic testing, which is preferred over the alternative approach of cyst content aspiration, as the latter carries the risk of leakage of cyst fluid with resultant dissemination of infection or

anaphylaxis. This most dreaded complication of echinococcal infection may lead to widespread and severe illness manifesting as fever, low blood pressure, shock, and possible death.

Medical treatment is often successful with mebendazole or albendazole (2). Ideally, following a course of perioperative antibiotics, surgical excision of the lesion is performed with the goal of removal of the parasite and the associated cystic lesions (5). Other treatment options include drug therapy alone or as a part of a special protocol known as PAIR (Percutaneous Aspiration, Infusion of scolical agents, and Reaspiration). The prognosis is generally good for patients responsive to oral therapy. Longitudinal sonographic studies are useful for monitoring the effects of treatment and chemotherapy (6). Prophylaxis is essential for complete eradication of the infection. Prevention can be achieved with sheep vaccination and medical treatment of infected canines.

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